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Case Report

Primary mediastinal large B-cell lymphoma with extensive cardiac involvement

Abstract

Data regarding the optimal therapeutic approach of patients with primary mediastinal B-cell lymphoma and cardia c involvement are limited. Reports suggest that multi-agent systemic chemotherapy, along with radiation therapy, may be effective in the treatment of these patients. However, a subgroup of patients do die suddenly due to myocardial rupture following the initiation of treatment. We report on the successful management of a patient with primary mediastinal large B-cell lymphoma with extensive cardiac involvement.

Introduction

A 61-year-old male with a past medical history of hypertension, dyslipidemia, presented to the emergency department complaining of dyspnea on exertion associated with palpitations for one week. A computerized tomography angiography of the chest obtained demonstrated extensive mediastinal lymphadenopathy with confluent soft tissue density narrowing and invading the central pulmonary arteries, coronary arteries, pericardium, right atrium, right ventricular free wall and left atrium (Figure 1). Transthoracic echocardiography showed a preserved systolic function with a heterogeneous mass embedded in the free wall of the right atrium extending into the free wall of the right ventricle, along with a 2.2cm×1.5cm mobile mass in the left atrium that was extending into the left ventricular free wall (Figure 2). A video assisted thoracoscopy, with biopsy, was performed which showed the mass to be a primary mediastinal B-cell lymphoma.

The patient was started on a dose-adjusted chemotherapy consisting of: etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin and rituximab, completing 6 cycles. Six months later, transthoracic echocardiography, showed near complete resolution of the cardiac mass; while an ^{18}F-fluorodeoxyglucose positron emission tomography with computed tomography scan showed activity in several mediastinal locations (Figure 3). Because of this, it was decided to continue treatment with adjuvant consolidative radiation therapy at the dose of 30 Gy to the mediastinum and 15-20 Gy to the area surrounding the heart. Nine months after initial diagnosis, repeat transthoracic echocardiography, computed tomographic angiography, and positron emission tomography with computed tomography showed complete remission of the disease (Figure 4). Two years after diagnosis, the patient is without evidence of active or recurrent disease.

Discussion

Primary mediastinal B-cell lymphoma accounts for 2%-4% of all non-Hodgkin's lymphoma [1]. It typically affects young adults, occurring at a median age of 35 years, with a female to male ratio of 1.7-2 to 1. The most common presentation is an anterior mediastinal mass which may invade local structures. Involvement of the heart is usually a late manifestation of disseminated lymphoma and is commonly asymptomatic [1,2].

The standard treatment includes a combination of chemotherapy and mediastinal radiation. However, routine mediastinal radiation in those patients with lymphomatous cardiac involvement has been associated with an increased risk of free wall rupture and increased incidence of ischemic heart disease [2]. Transthoracic echocardiography is considered the first diagnostic modality, being useful in the evaluation of possible pericardial effusions, valvular involvement, and left ventricular function. Computerized tomography and magnetic resonance imaging may aid in evaluating morphology, location, and extension of disease [3]. While ^{18}F-fluorodeoxyglucose

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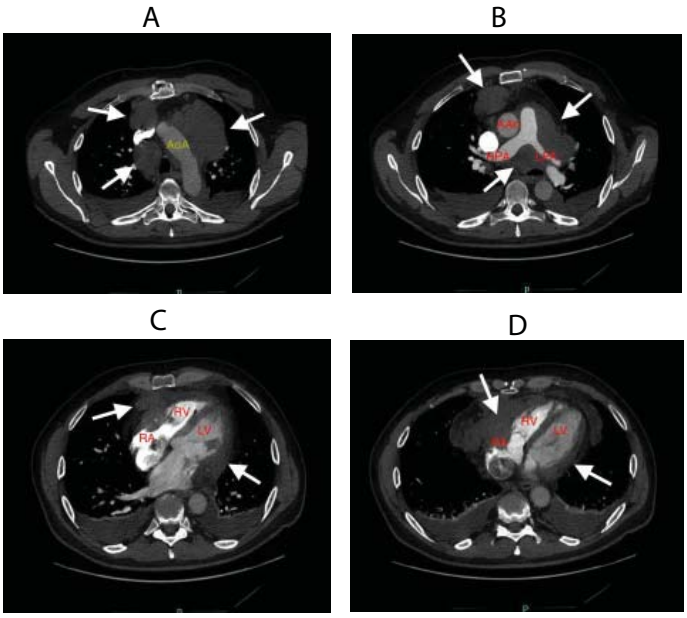

Figure 1: (A) Computerized Tomography Angiography of the Chest. Arrows demonstrate tumor invasion. Extensive and confluent multiple mediastinal lymphadenopathy. AoA: Aortic Arch.
(B) Computerized Tomography Angiography of the Chest. Arrows demonstrate tumor invasion. Confluent soft tissue density encasing and narrowing the main, right and left pulmonary arteries. RPA: Right Pulmonary Artery, LPA: Left Pulmonary Artery, AAo: Ascending Aorta.
(c) Computerized Tomography Angiography of the Chest. Arrows demonstrate tumor invasion. Soft tissue mass encasing and narrowing the coronary arteries, right atrium, right ventricular free wall, tricuspid annular plane, left atrial appendage and mitral valve plane. RA: Right Atrium, RV: Right Ventricle, LV: Left Ventricle.
(D) Computerized Tomography Angiography of the Chest. Arrows demonstrate tumor invasion. Soft tissue mass extending into the perivascular space of the anterior mediastinum measuring 6.2cmx8.2cmx8.2cm, with the inferior margin difficult to discern from the soft tissue mass invading the left heart border. Additionally noted is diffuse infiltration of the epicardium, pericardium and myocardial walls with encasement of the coronary arteries, which are diffusely narrowed. RA: Right Atrium, RV: Right Ventricle, LV: Left Ventricle.

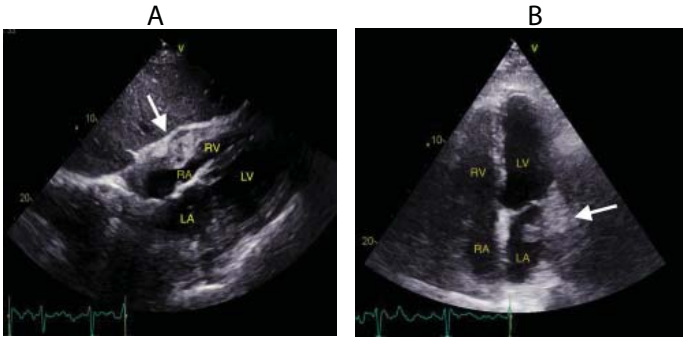

Figure 2: (A) Transthoracic Echocardiogram. Arrows demonstrate tumor invasion. Subcostal four-chamber view showing a heterogeneous mass embedded in the free wall of the right atrium extending into the free wall of the right ventricle. RA: Right Atrium, RV: Right Ventricle, LV: Left Ventricle.
(B) Transthoracic Echocardiogram. Arrows demonstrate tumor invasion. Apical four-chamber view showed a 2.2cmx1.5cm mobile mass in the left atrium that was extending into the left ventricular free wall. RA: Right Atrium, RV: Right Ventricle, LV: Left Ventricle.

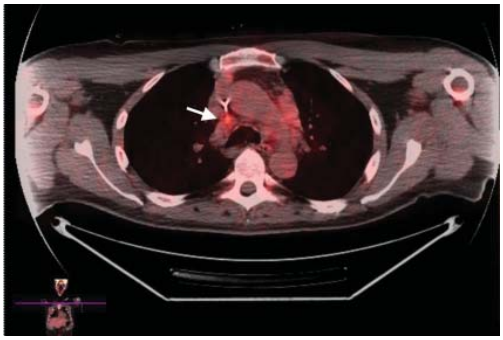

Figure 3: ^{18}F-fluorodeoxyglucose positron emission tomography with computed tomography scan showed activity in several mediastinal locations (white arrow).

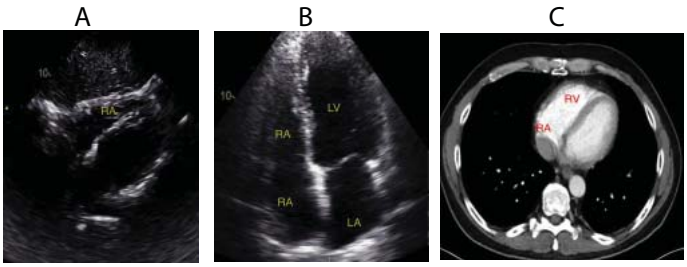

Figure 4: (A) Transthoracic Echocardiogram Subcostal showing complete resolution of the disease. Four-chamber view. RA: Right Atrium.
(B) Transthoracic Echocardiogram Subcostal showing complete resolution of the disease. Apical four-chamber view. RA: Right Atrium, RV: Right Ventricle, LA: Left Atrium, LV: Left Ventricle.
(C) Computed tomography angiography of the chest. RA: Right Atrium, RV: Right Ventricle.

positron emission tomography with computed tomography imaging is useful in the evaluation for the presence and extent of disease [4].

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